BRADYARRHYTHMIA IN MITRAL VALVE PROLAPSE TREATED WITH A PACEMAKER

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MITRAL valve prolapse receives increasing attention. This entity has many names, including systolic click-late systolic murmur syndrome, the floppy valve syndrome, and the billowing mitral leaflet syndrome.[3,4,5,7,12,19] It is an important cause of incapacitating chest pain with refractory arrhythmias.

Two patients with mitral valve prolapse presented with the manifestations of a sick sinus syndrome, with recurrent severe alternating brady and tachyarrhythmias. Both were treated with propranolol which increased the bradycardia and aggravated one patient's syncopal attacks. To overcome the latter, which may have resulted in a life-threatening seizure, a permanent transvenous pacemaker was implanted in both patients. Thereafter, propranolol was continued without ill effects and improvement of symptoms.

CASE 1

R. G., a 61-year-old white woman, was admitted to the University Hospital in 1976 because of chest pain, weakness, and syncope present for about two years. She was previously at another hospital where a diagnosis of mitral systolic click syndrome was made. A Holter electrocardiogram demonstrated a bradycardia of 48 to the minute, with one episode of ventricular coupling (Figure 1). She also demonstrated episodes of atrial tachycardia with recurrent sinus bradycardia and sinus arrest, and had several syncopal attacks. Her past history revealed a thyroidectomy for thyroid carcinoma in 1964, and a mastectomy in 1970. She was receiving 10 mg, of propranolol four times a day for her symptoms upon discharge from the hospital and continued without benefit.

Physical examination on admission demonstrated a thin, anxious woman complaining of chest pain and palpitations most of the day. The pain was

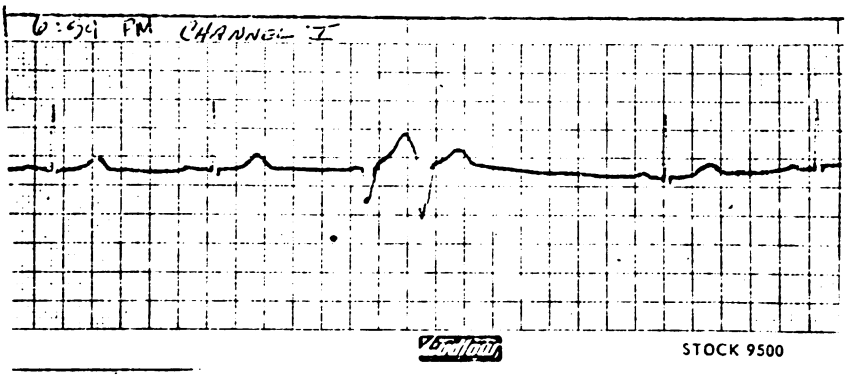

Fig. 1: Bradycardia with one episode of ventricular coupling

not typical of angina pectoris and neither radiated nor was related to effort or eating. It was dull in character, continuous, and not affected by nitroglycerin. Eyes, ears, nose, and throat were normal. The neck revealed an old thyroidectomy scar. The chest disclosed evidence of a left mastectomy and pes excavatum. The heart was not enlarged to percussion. The rhythm was of sinus origin and the rate was 43 to the minute. A systolic murmur was heard at the apex but no click could be detected. The blood pressure was 120/80 mm. Hg. The lungs were clear to percussion and auscultation. The abdomen revealed no masses and the extremities were free from edema or inflammation. The reflexes were normal.

A chest roentgenogram was within normal limits except for pes excavatum. An electrocardiogram showed bradycardia with a rate of 45, but otherwise was normal (Figure 2). Routine laboratory data were unremarkable.

An echocardiogram demonstrated a prolapse of the posterior leaflet of the mitral valve (Figure 3). On May 5, 1976 a 24-hour Holter tape electrocardiogram documented a bradycardia with occasional ectopic ventricular contractions and corresponding symptoms of palpitation and chest pain (Figure 4). The heart rate was 40 beats to the minute. Because the patient had bradycardia and persistent annoying symptoms with small doses of propranolol, a permanent transvenous pacemaker was inserted on May 11, 1976. Larger doses of propranolol then successfully controlled the tachyarrhythmias and chest pain without symptomatic bradyarrhythmia.

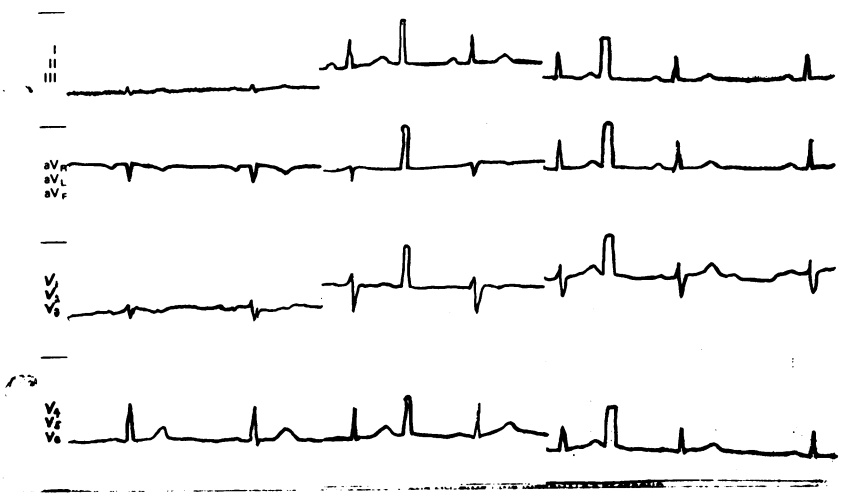

Fig. 2: Bradycardia with a rate of 45

CASE 2

D. M., a 53-year-old white woman, was admitted to the New York Infirmary in 1977 complaining of searing pain in the chest associated with severe palpitations. These symptoms recurred almost daily and were not related to effort, exertion, or food intake. Similar episodes had occurred about two years before, and had recently recurred with an increase in frequency and duration but without dyspnea or syncope. An electrocardiogram on several occasions revealed atrial flutter with a 2:1 and 3:1 conduction interspersed with periods of normal sinus rhythm, first degree atrioventricular block, and occasional long sinus pauses. For this she was treated with quinidine with very little success. Past history revealed diagnostic uterine curettage in 1969 and excision of a benign breast cyst in 1973.

On physical examination she appeared somewhat anxious and complained of chest pain. Eyes, ears, nose, and throat were normal. The neck was supple without palpable masses or venous distention. The chest was symmetrical and the expansion equal. Examination of the heart disclosed a loud, midsystolic click and high pitched late systolic murmur at the left sternal border and cardiac apex. The rhythm was of sinus origin with occasional episodes of flutter and sinus bradycardia with a rate of 33 beats to the minute. The blood pressure was 120/74 mm. Hg. The lungs were

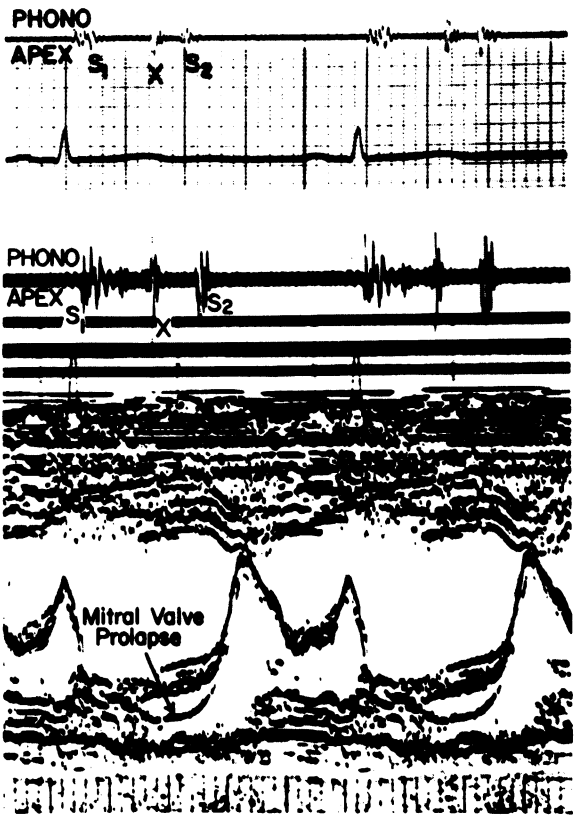

Fig. 3: Prolapse of the posterior leaflet of the mitral valve

clear to percussion and auscultation, and the rest of the physical examination was within normal limits. Routine laboratory data were normal, except for a mild hypochromic anemia, leukopenia, and thrombopenia attributed to quinidine therapy.

A phonocardiogram demonstrated a definite systolic click, and an echocardiogram revealed a prolapse of the posterior leaflet of the mitral valve (Figures 5 and 6). While being monitored in the coronary care unit, she had multiple episodes of atrial flutter and periods of sinus arrest (Figure 7). Quinidine was discontinued and she was given propranolol 10 mg. four times a day. With this regime she developed more periods of

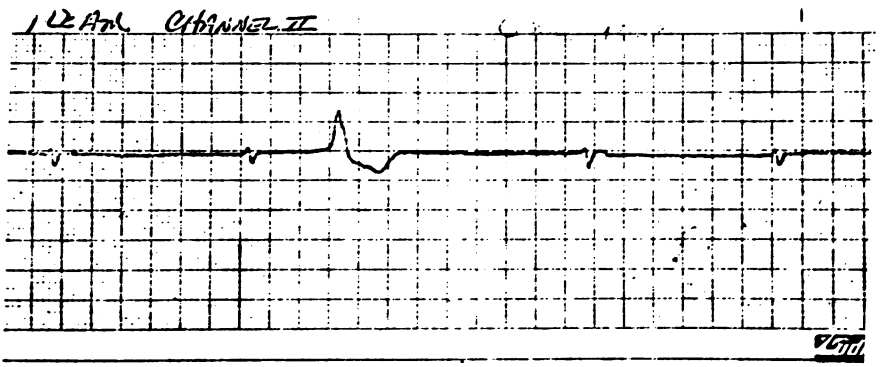

Fig. 4: A 24-hour Holter tape electrocardiogram showing a bradycardia with occasional ectopic ventricular contractions

sinus arrest and marked bradycardia. Because of this, a permanent transvenous pacemaker was implanted, and she was again given propranolol and digoxin with satisfactory control of all rhythm disturbances. She has remained free from chest pain and palpitations and has no bradycardia.

Discussion

Mitral valve prolapse was initially thought to be a relatively rare, benign extracardiac phenomenon, but recently has emerged as an extremely common disorder, not always benign.[12] Its cause is uncertain and it is manifested by posterior displacement of one or both mitral leaflets across the atrioventricular groove into the left atrium during systole. The actual prevalence of this syndrome is at present unknown, but some reports indicate that it is a common echocardiographic finding in presumably healthy individuals. Markiewicz et al. in 1976[9] reported an incidence of 17% in 100 presumably healthy young women. Procacci et al.[10] demonstrated mitral valve prolapse in 6.3% of 1,169 young, healthy women. Other observers have found this entity in young, asymptomatic subjects in appreciable numbers but less frequently in men.[12]

The clinical spectrum varies from a nonejection systolic click with or without a late systolic murmur and no mitral insufficiency to severe mitral incompetence with a holosystolic murmur and no click. The click, usually midsystolic, may be heard loudest at the cardiac apex, but can also be detected at the left sternal border or midprecordium. At times the click may be multiple, and at other times disappear entirely because the entity is

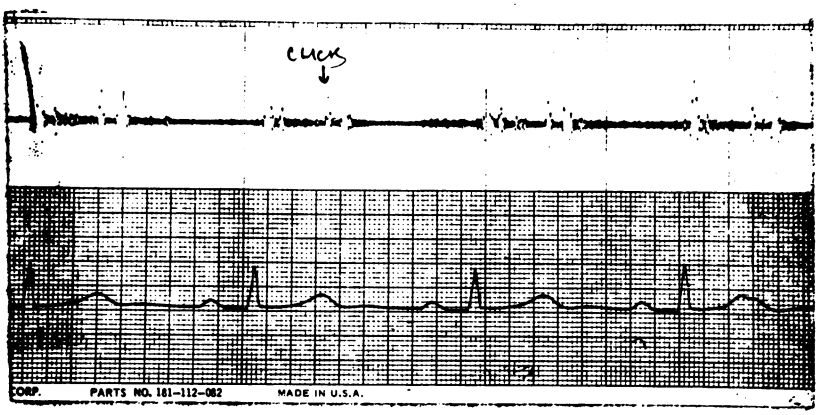

Fig. 5: Phonocardiogram showing a systolic click

a dynamic one. The murmur is usually heard in late systole when present, and begins after a silent interval following the first sound. It may be introduced by a loud click and it often has a musical or "whooping" character. Various body positions and maneuvers augment the murmur.[12,19] Passive leg raising increases venous return and increases the left ventricular diameter, delaying the murmur and click until later in systole. Standing decreases venous return and causes the murmur to lengthen and become louder. Squatting causes a simultaneous increase in venous return which increases ventricular size, delaying and shortening the murmur. The timing of the click and murmur is influenced by other specific manipulations, i.e., the Valsalva maneuver, exercise, and amylnitrate inhalation which also diminishes ventricular size, causing an early appearance of the murmur and click, whereas vasopressors delay the onset of both.[18]

The major clinical problems of the mitral valve prolapse syndrome consist of chest pain, palpitations, dyspnea and weakness, and arrhythmias. Chest pain is very common and usually atypical, unrelated to effort, stress, cold, food, or nitroglycerin. It also differs from true angina in that it lasts longer and does not radiate in the usual manner. Palpitations disturb the patient, often from a tachyarrhythmia, but more often they are unexplainable. Dyspnea and weakness may be manifestations of cryptogenic abnormalities of left ventricular function or purely related to some as yet unrecognized neuropsychiatric disorder.[7,17] Many patients with the systolic click-late systolic murmur syndrome have minor bony abnormalities similar to those found in Marfan's syndrome, such as pes excavatum, straight spine, and scoliosis.[14,15]

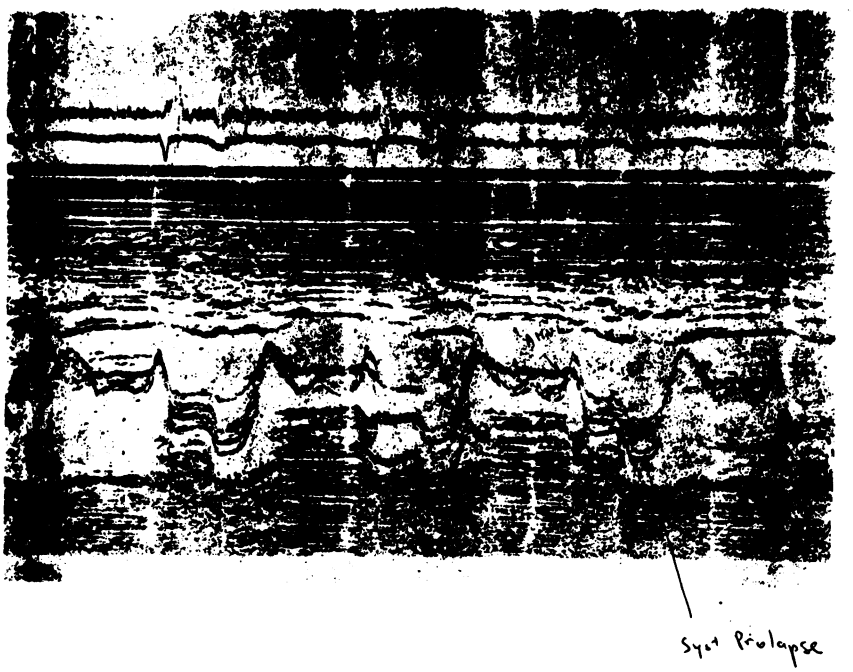

Fig. 6: Echocardiogram demonstrating prolapse of the posterior leaflet of the mitral valve

About 25% of patients with this syndrome are entirely asymptomatic and the diagnosis rests on echocardiography which has contributed considerably to understanding the entity of mitral valve prolapse. Late systolic prolapse of one or both leaflets can be directly seen by echocardiography as posterior movement which interrupts the normal anterior motion. The click occurs during maximum leaflet prolapse and is best demonstrated by phonocardiography. Combined echocardiography and phonocardiography clinch the diagnosis.

Arrhythmias are an important component of the mitral valve prolapse syndrome. They are not always apparent on a routine electrocardiogram but continuous ambulatory monitoring by the Holter technique is often rewarding. Exercise alone may bring on attacks. Occasionally, an arrhythmia in an otherwise healthy heart may give a clue to the presence of the mitral valve prolapse. The most common arrhythmias are premature ventricular beats, tachyarrhythmias, bradyarrhythmias, and occasionally sinus arrest. Gulotta et al.[20] reported three patients with mitral valve prolapse who developed different degrees of heart block, two of whom

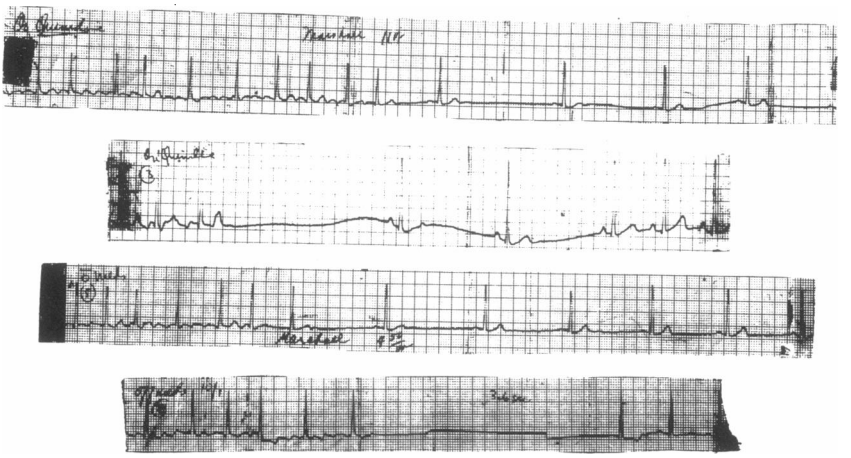

Fig. 7: Multiple episodes of atrial flutter and periods of sinus arrest

received permanent transvenous pacemakers. It is not certain whether these cases represent coincidence or an unusual variation in the mitral valve prolapse syndrome. These cases differ from ours inasmuch as the installation of the pacemaker was not related to propranolol.

Swartz et al.[21] in 1977 reviewed 589 patients with mitral valve prolapse who had an arrhythmia. He found premature atrial ventricular contractions or both in 55%, premature ventricular contractions in 45%, supraventricular tachycardia in 6.1%, and ventricular tachycardia in 6.3%.

De Maria et al.[22] described continuous Holter monitoring in 20 patients with mitral valve prolapse. Several of these patients had multiple premature contractions, either paired or multifocal, and they noted that six of the 20 had episodes of abrupt sinus bradycardia, sinus arrest, or sinus arrhythmias. Atrial extrasystoles or unexplained sinus tachycardia occurred in an additional four patients. Both of our patients presented similar findings. Criley and his coworkers[24] also monitored 17 patients with mitral valve prolapse for periods of 10 to 168 hours and found 15 with either atrial or ventricular arrhythmias. De Silva and Shubrooks in 1977[23] described a 20-year-old boy with documented mitral valve prolapse, sinus bradycardia with marked sinus arrhythmia, and a high-grade atrioventricular block of many years duration, probably beginning in infancy consistent with a congenital etiology.

The etiology of the arrythmias with mitral valve prolapse remains

unknown. Criley et al.[24] suggested two mechanisms by which mitral dysfunction and ectopic beat formation may be related: mechanical stimulus of the left ventricle or atrium from excessive movement of the distended, blood-laden valve leaflet and ectopic impulse formation from stretching of the valve leaflet or myocardium. Witt et al.[25] have demonstrated that the mitral valve can act as a site of ectopic impulse formation *in vitro*. Such other mechanisms as a primary myocardial disorder or an abnormality in left ventricular contractility have been suggested as the basis of the arrhythmias associated with mitral valve prolapse.

Leichtman et al. in 1976[13] reported three patients with mitral valve prolapse and episodes of abrupt sinus bradycardia, exaggerated sinus arrhythmias, and sinus arrest. Two of these patients who developed syncope were treated by pacemaker to avoid sudden death. Their groups were members of a family of 11 with a high preponderance of mitral prolapse and sinus bradycardia. They suggest that excessive vagal tone was responsible for both the bradycardia and sinus arrest. Their patients also differed from ours inasmuch as the sinus arrest and syncope occurred without propranolol.

It is generally agreed that the treatment of choice for the mitral valve prolapse syndrome is propranolol, which usually controls both the chest pain and tachyarrhythmia. However, when bradycardia and sinus arrest develop during propranolol therapy, it becomes dangerous to use this drug. We have overcome this dilemma by a permanent transvenous pacemaker and administering propranolol in the proper dosage to control pain and dysrhythmia. We believe that we are the first to employ such a combination.

SUMMARY

Mitral valve prolapse is now known to be a fairly common syndrome. It is an important cause of incapacitating chest pain and refractory arrhythmias. Recently we encountered two patients with mitral valve prolapse who presented with a sick sinus syndrome with recurrent and alternating brady and tachyarrhythmia of a severe form. Both were treated with propranalol, which increased the bradycardia. In order to be able to continue therapy with the latter we had a transvenous pacemaker implanted in both patients. Thereafter propranalol was continued without ill effects and improvement of symptoms.

ACKNOWLEDGMENTS

We thank Drs. Joseph Cunningham and Louis Delli Pizzi for their assistance in the installation of the pacemakers.

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